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Our data reveal several interesting observations. Firstly, skin tumours that are strongly associated with specific gene disorders can be used to detect respective patients with genetic conditions from electronic health records. A related approach has been described, where diagnostic coding data are refined using multiple electronic records such as prescription data to improve identification of patients with diabetes.<sup>4,5</sup> Patients with CCS in this series who had not been clinically diagnosed stand to benefit from genetic testing, counselling and skin surveillance. Secondly, the presence of associated tumours in CCS such as salivary gland tumours<sup>6</sup> and malignant transformation<sup>2</sup> can be monitored.

The generalizability of this approach to other genetic disorders beyond CCS, where cancer and skin appendageal tumours may occur in combination, is important. Muir–Torre syndrome, where sebaceous carcinoma and bowel cancer develop, Birt–Hogg–Dubé syndrome, where trichodiscomas and renal cell carcinoma present, and Reed syndrome, where leiomyomas and renal cell carcinoma present, are pertinent examples. In these conditions, screening for malignancy could be initiated in relevant individuals, and in the case of Muir–Torre syndrome chemoprevention with aspirin can reduce cancer risk.<sup>7</sup>

Recently in the UK, national registration has been initiated for skin cancer.<sup>8</sup> As we transition to a digital health service, it is conceivable that all skin pathology cases will be searchable, including skin appendageal tumours. We propose that the application of filtering algorithms could be applied to increasingly comprehensive electronic health records, allowing patients with an underlying genetic condition to be detected. Limitations of this proof-of-principle study are that some patients with CCS will have only one biopsy at the time of study, and typographical errors in free-text coding can result in omission of patients. Our method found that most patients with potential CCS identified from electronic pathology records during this interval had a confirmed diagnosis of CCS. Future studies, as genetic testing for CCS becomes established as a standard of care, should also investigate the number of patients with a confirmed diagnosis of CCS who are not identified using our method in the electronic pathology records. Nonetheless, these data are relevant, at a time when artificial-intelligence-based analysis of large datasets such as national cancer registries is underway.

The ability to detect any potential genetic disease by proxy methods raises issues regarding consent, similarly to those raised in the context of conventional genetic testing. Public perception of genetic testing is changing, and patient group engagement will be important in guiding how such algorithms may be acted upon when they detect a patient with a genetic predisposition. For example, it may involve notification only when a helpful intervention is possible. It would be beneficial to conduct qualitative interviews in individuals with conditions such as CCS to capture their perceptions of the risks and benefits of being diagnosed in this manner. Patients have the right not to know about a genetic diagnosis, yet some may consider it a failing of a digital healthcare system if data point to a genetic diagnosis where screening or chemoprevention is possible, and yet no action is taken. We highlight that electronic

health records are a rich resource, and continue to offer new facets that can impact on patient care.

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## The prevalence of hidradenitis suppurativa is shown by the Secure Anonymised Information Linkage (SAIL) Databank to be one per cent of the population of Wales

DOI: 10.1111/bjd.19210

DEAR EDITOR, Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease with multiple inflammatory skin lesions

('boils') in flexural locations.<sup>1</sup> Prevalence of HS remains controversial<sup>2,3</sup> and the study aim was to search the Secure Anonymised Information Linkage (SAIL) Databank to identify diagnosed and undiagnosed HS cases in Wales, UK.

The SAIL Databank provides access to routinely collected primary care data, linked to Patient Episode Database for Wales (PEDW) data, covering 2.5 million individuals within the Welsh population of just over 3 million people.<sup>4</sup> The prevalence of diagnosed cases of HS was determined using Read Codes M25y1 and M25y111. Undiagnosed 'proxy' cases were identified using the skin boil algorithms developed and validated for the UK Clinical Practice Research Datalink (CPRD).<sup>3</sup> The algorithms require a record of at least five skin boils, on separate occasions within the primary care databank. Validation in CPRD makes use of General Practitioner (GP) questionnaires, which are limited by GPs having several thousand patients under their care, and so GPs themselves are often reliant on the electronic record. While SAIL does not have access to GP validation, it is linked to HealthWise Wales (HWW), a register of people willing to take part in research and return health questionnaires.<sup>5</sup>

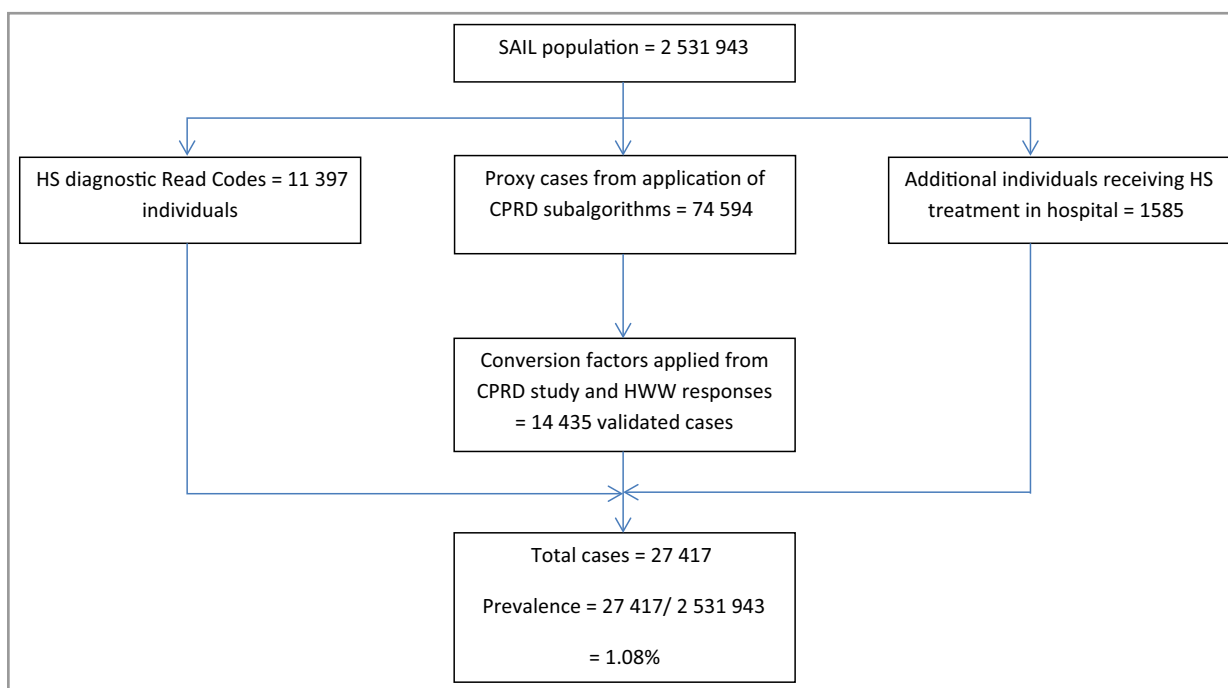
An online questionnaire was sent to HWW participants to identify outbreaks of skin 'boils' during the previous 6 months in flexural locations and a minimum of two boils. The questionnaire has a published specificity of 97% and a sensitivity of 90% in identifying people with HS.<sup>6</sup> Of the 1481 SAIL participants who answered the questionnaire, 86 (5.8%) gave responses consistent with HS. Five of these individuals had a HS Read Code in SAIL and 14 were identified by the CPRD algorithms. Manually checking a representative sample of the other 67 positive

respondents, there were no relevant consultations, such as for skin boils or pilonidal sinus, recorded in SAIL. Due to a lack of data, validation of CPRD algorithms was possible for subalgorithm 2(b) only, for which eight HWW respondents gave positive responses and 31 were negative for HS (21% positive rate). As a consequence, validation data were taken from the CPRD study for the other subalgorithms.<sup>3</sup>

From the SAIL population of 2 531 943 individuals, 11 397 had an HS diagnosis in SAIL up to 30 June 2017. A further 1585 SAIL patients were treated for HS in secondary care as identified by PEDW data. Hence, the point prevalence of diagnosed HS in the Welsh population was 5.1 per 1000, or 0.51%. Application of the CPRD hierarchical subalgorithms identified 74 594 additional proxy cases, reduced to 14 435 criteria-diagnosed cases after validation, using the conversion factors developed in the CPRD study<sup>3</sup> and the HWW-validated conversion factor for subalgorithm 2(b). Overall HS prevalence is therefore 10.8 per 1000, or 1.08% (Figure 1).

Of the 100 HS patients attending a specialist secondary care HS clinic in Wales, 85 of the cohort were present in SAIL, 72 as diagnosed cases and a further nine within the subalgorithms, with four cases not captured. This confirms relatively high capture of secondary care HS cases by SAIL, with 85% correctly transcribed into the primary care record and only 5% missed by either the diagnostic code or a subalgorithm.

During the period from 2000 to 2016, the number of newly diagnosed HS cases per year increased nearly fourfold, from 200 to 765, likely owing to increasing recognition of HS. The female-to-male ratio of newly diagnosed cases was 3 : 1 and peak age of diagnosis was in the third decade of life.






**Figure 1** Data sources and their contributions to the overall hidradenitis suppurativa (HS) prevalence figure and validation process. SAIL, Secure Anonymised Information Linkage Databank; CPRD, Clinical Practice Research Datalink; HWW, HealthWise Wales.

Limitations of the study include a relatively short time frame for HWW data collection, which was only able to validate subalgorithm 2(b) in this cohort. However, CPRD covers Wales, as well as the English population, and so validation data for CPRD will also be valid for the Welsh population. The 5.8% rate of positive returns from the HWW questionnaire is high and indicates a degree of case ascertainment bias. Lack of relevant SAIL consultations in most of this group suggests that either the individuals had mild disease or they chose not to see their GP, or both. Our prevalence figure may be an underestimate because HWW data were used only as a validation tool for SAIL data.

Comparing the SAIL results with prevalence figures from CPRD, there is quite close agreement. Prevalence of diagnosed cases in CPRD was 5.4 per 1000 (0.54%).<sup>3</sup> Prevalence of HS in CPRD rose to 0.77% when validated proxy cases were included, and 1.19% when probable cases with a history of 1–4 flexural skin boils on separate occasions were included.<sup>3</sup> The SAIL prevalence of 1.08% adds further weight to the evidence that HS is a relatively common condition in the UK.

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**Conflicts of interest:** J.R.I. is Editor-in-Chief of the *BJD*. He is a consultant for UCB Pharma, Novartis, Viela Bio and Kymera Therapeutics; he received travel expenses and a speaker's honorarium from UCB Pharma. The remaining authors declare they have no conflicts of interest.

## Acral lentiginous melanoma: clinicopathological characteristics and survival outcomes in the US National Cancer Database 2004–2016

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DEAR EDITOR, Acral lentiginous melanoma (ALM) is one of four subtypes of cutaneous melanoma (CM), comprising 2–3% of all melanomas in the USA.<sup>1</sup> It occurs predominantly on the palms, soles and nail beds. It is a common subtype found in people with darker skin types. ALMs are generally associated with a poor prognosis.<sup>2</sup> Understanding of the demographics, outcomes and treatment of this type of melanoma can be aided by studying a larger cancer registry.<sup>3</sup> This study, to our knowledge the largest retrospective study of ALM, aims to take a closer look at the difference in demographics and overall survival (OS) of patients with ALM.

The National Cancer Database (NCDB) was queried for all patients with ALM from 2004 to 2016. The NCDB gathers information from more than 1500 accredited cancer facilities in the USA and Puerto Rico. It collects approximately 70% of cancer diagnoses in the USA annually.<sup>4</sup> To reduce ambiguity, only patients with histologically confirmed cases of ALM as the primary malignancy were included. Only cases in the upper extremities, including the palms and fingernails, and in the lower extremities, including the soles and toenails, were included. Univariate relationships between overall mortality from ALM and known and suspected prognostic variables were assessed using the Kaplan–Meier (KM) method. Variables that showed significant differences ( $P < 0.05$ ) on univariate KM analysis, including age, sex, race, Charlson/Deyo comorbidity (CDCC) index, stage, ulceration, Breslow thickness, income status, facility type, primary site, insurance status and treatment type, were included in the Cox regression. As this study